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A case of recurrent eccrine porocarcinoma

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Case Report

Abstract

BACKGROUND: Eccrine porocarcinoma, first described in 1963, is a rare malignant lesion arising from the eccrine sweat glands. It is usually a primary tumor, even more commonly, a malignant degeneration of an eccrine poroma. It usually affects elderly, and is located commonly in the lower extremities. About 20% of the cases will reappear after treatment. The treatment consists of wide local excision of the primary lesion. This uncommon skin tumor is locally aggressive and has a high recurrence rate.

CASE REPORT: We report a 95-year-old woman with a large tumor on the left knee. A large number of tumoral cells showed immunolabeling of p63 supporting squamous or myoepithelial differentiation. They were also positive for cytokeratin-7 (CK7), epithelial membrane antigen (EMA), and carcinoembryonic antigen (CEA) highlight luminal differentiation in some of the vacuolated cells and this was also seen with periodic acid-Schiff-diastase (PAS-diastase) staining.

CONCLUSION: Eccrine porocarcinoma is a rare aggressive form of skin cancer with unknown etiology, and little guidance is available in the literature on exact protocols for treatment and follow up. It should be on the differential diagnosis of any suspicious skin lesion seen by the plastic surgeon. Histologic assessment is indicated in suspicious lesions.

KEYWORDS: Eccrine Porocarcinoma, Recurrence, Sweat Glands

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Introduction

Eccrine porocarcinoma (EPC) is a rare malignant cutaneous appendageal tumor that develops from intraepidermal eccrine sweat duct.[1] Pinkus and Mehregan were the first to describe EPC in 1963.[2] 0.005% to 0.01% of all epidermal skin neoplasms were caused by these tumors.[3] Patients' age ranging is being reported from 21 to 90 years old. It occurs predominantly in elderly people who are more than 60 years, and indicates a female predominance. Its etiology is not well understood. However, some studies have demonstrated that the tumor developed from a pre-existing eccrine poroma.[4]

From another point of view, prompting factors include chronic light exposure, exposure to chemical agents, and immunosuppression. Porocarcinoma is a hazardous disease because of high rate of recurrence after resection and aggressive behavior.[5] Due to seldom occurrence of the problem, there is controversy regarding both presentation and management. One characteristic histologic feature of EPC is ductal differentiation of poromatous basaloid epithelial cells that forms an irregular tumor shap.[6] Prevalent occurring sites consist of lower extremities, followed by the trunk, head, and upper and lower limbs.[7]

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Case Report

She was a 95-year-old woman with a large tumor on the left knee. The sections of biopsy in year 2014 showed skin and underlying tissues containing a highly infiltrative adnexal carcinoma. The tumor composed predominantly of nests of epithelial cell, with round nuclei, and vacuolated cytoplasm lying myxoid stroma. Some cells had cytoplasmic vacuoles. Many of the nests showed central zone necrosis. Many mitotic figures were presented. Areas of squamous differentiation were also accompanied by focal differentiation to form ductal lumina. These areas mostly consisted of porocarcinoma cells. In some areas of the tumor, there was abundant myxoid stroma; that was less expected in porocarcinoma, and suggested chondroid syrindoma. The neoplasm had high infiltrative growth pattern, extending to all of the margins of the biopsy (Figure 1).

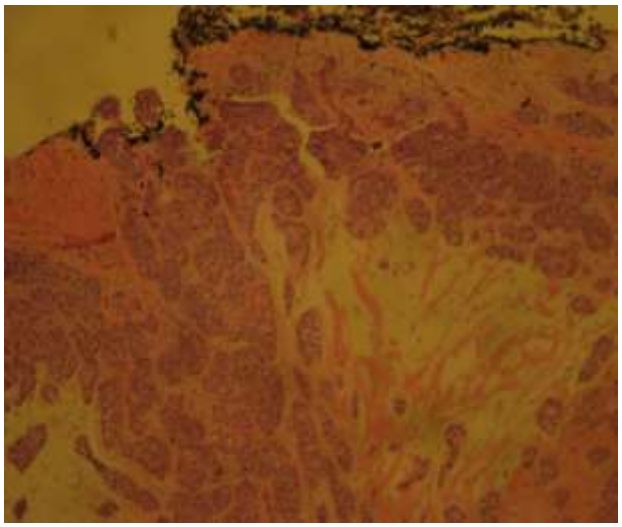

Figure 1. Hematoxylin and eosin (H&E) staining (× 100) showing high infiltrating growth pattern with margin involvement

There was also pagetoid invasion of overlying epidermis. Most of the tumoral cells showed immunolabeling for p63 supporting squamous or myoepithelial differentiation (Figure 2).

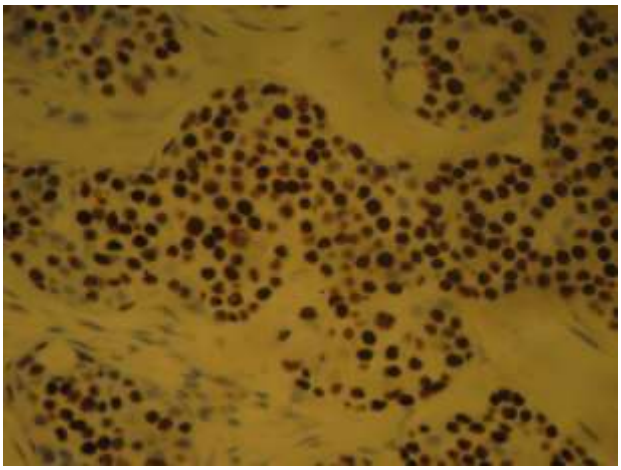

Figure 2. P63 staining (× 400) strongly supporting squamous differentiation

Most of the cells were also positive for cytokeratin-7 (CK7), epithelial membrane antigen (EMA), and carcinoembryonic antigen (CEA) highlight luminal differentiation (Figure 3).

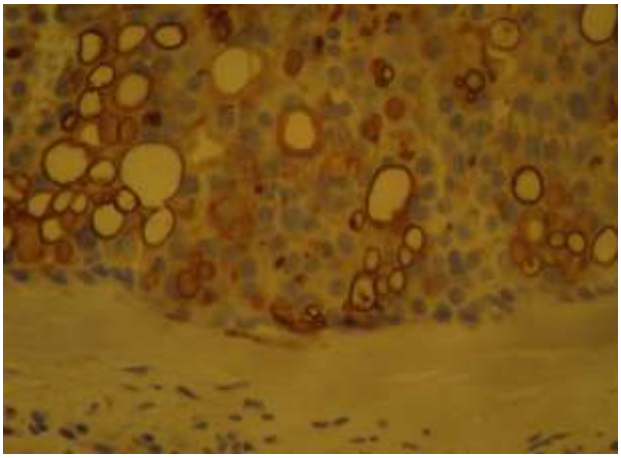

Figure 3. Immunohistochemistry (IHC) staining (× 400) of carcinoembryonic antigen (CEA) supporting luminal differentiation

A few number of the vacuolated cells were also seen via periodic acid-Schiff-diastase (PAS-diastase) staining (Figure 4).

There was a few amount of intracytoplasmic mucins indicated in some of the cells. Sections of excision in year 2017 showed a recurrence of the tumor that had the identical morphology to the one seen in year 2014. Extensive areas of necrosis were seen.

The recurrent tumor appeared to have been excised incompletely again.

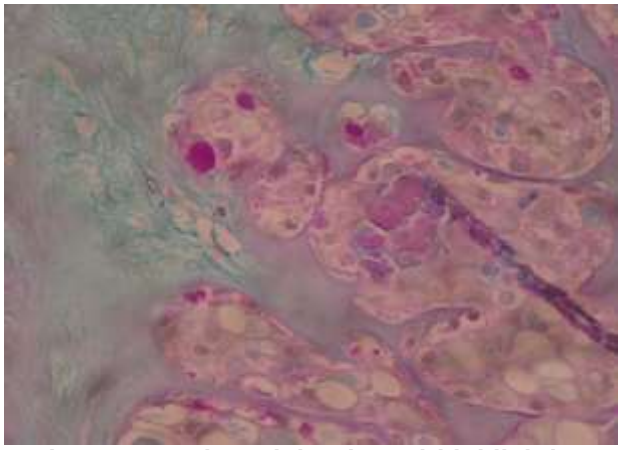

Figure 4. Mucin staining (× 400) highlighting luminal differentiation

Discussion

Theoretically, EPC develops from benign to malignant. EPC is a biologically aggressive neoplasm and has a high rate of recurrence and metastasis.[8] Its histogenesis is yet to be known, but is believed to have an origin from the acrosyringium.[9,10] From the view of histology, EPC has two types: intraepidermal, and dermal porocarcinoma. The intraepidermal type develops horizontally and generates pagetoid infiltration along the epidermis; the dermal form indicates nodular aggregates, usually without attachment to the epidermis.[11]

Our patient had an intraepidermal lesion presented with mitotic figures, such as affected dermis. The EPC clinical manifestations are not specific, and different types of papule, nodule, and plaque form lesions with various sizes (1-10 cm) could be seen.[12-14] According to Mulinari-Brenner et al. review article, the lesions can be sized between 1.2 × 2.0 and 4 × 5 cm.[15] EPC is more common in women and has had higher incidence in the age group of 70 years in some case studies.[15,16]

Bowen's disease, extramammary Paget's disease, cutaneous metastasis, amelanotic melanoma, cutaneous lymphoma, cutaneous squamous cell carcinoma, and other primary skin appendageal tumors should be in mind as differential diagnosis.[16] Robson et al. in a case series of 69 patients with EPC concluded that aggressive behavior might be the exception; as they reported 17, 10, and 20 percent that local recurrence, distant metastasis, and lymph node metastasis in EPC cases, respectively.[17] This issue is confirmed by some other researchers.[8-10,18]

It should be noticed that despite these case series, diagnosis, treatment, and prognosis issues of EPC are still provocative due to the lack of enough data on this disease. Some authors recommended the four histopathological findings as predictive factors in patients with EPC;[17-19] as lymphovascular invasion, and invasion depth of more than 7 mm or more than 14 mitoses per 10 high-power field (HPF) was associated with death, and infiltrative growth pattern was prognostic of local recurrence.[17,19]

Conflict of Interests

Authors have no conflict of interests.

Acknowledgments

None.

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